Table S1—Key studies using patient-reported measures of narcolepsy.

Scale	Study Authors, Date[a]	N	Population[b]	Study Objectives: Outcomes
CETQ	Moore WR, et al, 2007[46]	356	78 patients with NC vs 78 controls with OSA; mean age (range): 53.5 (20-84) years (y); 58% women	Pilot validation study: ESS sensitivity/specificity values were 0.94/0.99 and 0.90/0.99 for questions 1 and 2, and ranged from 0.74/0.99 to 0.87 0.94 for questions 2-5; PPV/NPV values ranged from 0.75/0.99 to 1.00/0.99 for all 5 questions
ESS[c]	Johns MW, 1991[36]	180	150 adults with various sleep disorders including 13 with narcolepsy and 30 normal controls; mean (±SD) age: 46.6 (12) y; 61.5% men	Initial/introductory study: ESS scores were significantly higher (indicating ES) in participants with narcolepsy vs controls or primary snorers (P<0.001 for both); ESS was significantly correlated with nocturnal sleep latency (r = -0.379 [n=138], P<0.001) and daytime sleep latency (MSLT; r = -0.514 [n=27], P<0.01)
	Johns MW, 2000[37]	530	Men and women with narcolepsy participating in 4 modafinil treatment trials (N=530), aged 17 to 68 y; sex percentages NR	To review and summarize data from large clinical trials of modafinil for narcolepsy symptoms: ESS sensitivity/specificity was 93.5/100.0 (cut-off > 10); ROC curves showed greater specificity of ESS over MSLT and MWT
	Parkes JD et al, 1998[38]	371	188 patients with narcolepsy, 62 with hypersomnia, and 10 with OSA vs 188 normal controls; mean age (range): 54.3 (12-85) y; 51.9% women	To use ESS for assessment of daytime sleepiness among other scales to develop a new measure for narcolepsy: ESS scores were 5 times higher in patients with narcolepsy vs controls (mean [SD] ESS score in narcolepsy patients: 19.6 [± 3.0], median [range]: 20 [9-24]); ESS sensitivity/specificity was 97%/100% (cutoff of 15)
	Frauscher B et al, 2013[39]	100	Adults with narcolepsy, 87 with NC, 13 without cataplexy (N); median age (range): 39 (16-78) y; 56% men	Chart review of consecutive patients: Median (range) ESS score was 18 (10-24); ESS scores tended to be higher in NC vs N patients

	Kendzerska TB et al, 2014[40]	NR	Patients across 35 studies with a variety of sleep disorders and normal controls; mean/median age, sex percentages NR	To summarize study data on psychometric properties of ESS via review of 35 published studies: Cronbach's alpha (internal consistency) ranged from 0.73-0.86; test-retest correlation was 0.82 in one good-quality study; pooled correlations for construct validity were moderate for MWT (-0.43) and weak for MSLT (-0.27); differences in scores between patients with narcolepsy and controls were clinically meaningful in multiple studies
NSAQ	Mamelak et al, 2004[d56]	25	Adults with NC (N=25); mean (±SD) age: 52.6 (8.8); 72.0% women	A pilot study to assess the effect of nocturnal, open-label sodium oxybate treatment for 10 weeks on sleep architecture in patients with NC: patients reported dose-related, subjective improvements in NSAQ measures including daytime sleepiness (76%), nighttime sleep quality (81%), ability to concentrate (67%), and overall condition (81%); other, objective measures used in the trial also showed improvements
	Mamelak et al, 2015[d54]	202	Adults with NC (N=202) either naïve to sodium oxybate or not previously titrated with it to clinical effect; mean (±SD) age: 41.9 (14.9); 65% women	To evaluate the efficacy of long-term (12- weeks) open-label treatment with sodium oxybate for reducing cataplexy attacks and improving excessive sleepiness using the NSAQ as the primary measure: percentages of patients reporting improvements in NSAQ items at Week 6 of treatment ranged from 87% (number cataplexy attacks) to 61% (number sleep paralysis episodes), and these improvements were maintained through Week 12
NSSQ	Mitler MM et al, 1982[52]	18	10 patients with narcolepsy and 8 normal controls matched for age; mean age; 43.8 y; 50% women	Initial use to determine effect of treatment for ES and correlation with MWT: significant differences post-treatment vs baseline were observed for all 7 items of scale, and sleep attacks, but not sleepiness; changes were consistent with improvements in MWT

	Rogers AE, 2001[53]	29	Adults with narcolepsy (with/without cataplexy) all being treated with stimulants and for cataplexy when present; mean (range) age: 43.7 (18-64) y (N=29); 58.6% women	To compare treatment including scheduled sleep periods + stimulant medications vs stimulant medications alone: correlations of NSSQ with other ES measures were inconsistent; NSSQ baseline severity scores correlated highly with post-treatment improvements (*P*=0.006); pretrial analyses also showed reliability of NSSQ items, sleepiness, sleep attacks, and cataplexy (*r*s =0.91, 0.83, and 0.68, respectively).
SNS	Sturzenegger C et al, 2004[15]	153	57 patients with definite NC (n=41) or narcolepsy with possible cataplexy (NpC; n=16), 56 patients with NNH, and 40 normal controls; median (±SD) age: 46 (18) y; 58% men	To evaluate clinical characteristics of narcolepsy, and assess a new narcolepsy scale, the SNS, in comparison with other, established scales (UNS, ESS): SNS scores were significantly different in all narcolepsy (NC + NpC) vs NNH patients (*P* < 0.001), and between both NC and NpC vs NNH (*P* < 0.046 for NpC vs NNH); SNS had sensitivity/specificity for narcolepsy of 96%/98% vs 98%/56% for the UNS.
	Sturzenegger C et al, 2006[49]	175	33 patients with NC, 142 with ES of other origins (e.g., SDB, IH); mean/median age, sex percentages NR	To compare SNS sensitivity/specificity with that of UNS and ESS to diagnose narcolepsy and identify hypocretin-1-deficient narcolepsy: SNS sensitivity/specificity for narcolepsy diagnosis was 85%/92% (cutoff < 0) vs 100%/77% for UNS (cutoff ≥14) and 81%/73% for ESS (cutoff ≥14); sensitivity/specificity for identification of hypocretin-1-deficient patients was 93%/92% for SNS vs 100%/77% for UNS and 92%/73% for ESS.

	Sturzenegger C et al, 2014[48]	207	85 patients with NC, 122 with ES of other origins (e.g., IH, SDB); mean/median age, sex percentages NR	To compare SNS sensitivity/specificity with that of UNS and ESS to diagnose narcolepsy and identify hypocretin-1-deficient narcolepsy: SNS sensitivity/specificity for narcolepsy diagnosis was 86%/89% (cutoff <0) vs 100%/62% for UNS (cutoff ≥14), and 91%/54% for ESS (cutoff ≥14); for identification of hypocretin-1-deficient narcolepsy, sensitivity/specificity was 92%/89% for SNS vs 100%/62% for UNS and 93%/54% for ESS
SSI	Anic-Labat, 1999[26]	983	63 patients with clear-cut NC and 920 with other sleep disorders entering the Stanford Sleep Disorder Clinic; mean (±SD) age: 46.2 (2.1); 56% women	To validate the SSI section focused on cataplexy for diagnosis of cataplexy: ROC analysis showed that occurrence of cataplexy (i.e., muscle weakness) triggered by hearing and telling a joke (risk = 73.3% "yes" vs 1.7% "no") and anger (risk = 91.7% "yes" vs 45.8% "no") were the most important predictive factors for diagnosis of cataplexy and differentiation from other sleep disorders; laughter was the second most useful discriminator if patients denied hearing or telling a joke as a trigger of cataplexy symptoms (risk = 32.5% "yes" vs 0.6% "no")
UNS	Hublin C et al, 1994[44]	488	24 patients with NC, compared with 7 disease groups: OSA (n=29); multiple sclerosis (n=25): depression (n=79); epilepsy (n=116); sciatica (n=105); alcohol abuse (n=38); and neurovegetative symptoms (n=72); mean (range) age: 42.8 (17-71) y; 50% women	To validate the UNS for NC: the mean UNS score was 27.3 in patients with NC vs 9.6 in the OSA group and between 5 and 6 in the other comparison groups; sensitivity/specificity for NC was 98.8%/100% (cutoff ≥14)

	Wing YK et al, 2000[45]	234	17 patients with narcolepsy, 196 with various other sleep disorders, and 21 controls; mean (±SD) age 38.0 (17.5) y; 64.7% male	To validate the Chinese version of the UNS (CUNS) for narcolepsy: Mean (± SD) CUNS scores were significantly higher in patients with narcolepsy vs all other groups (18.6 [4.7]; 95% CI: 16.2-21.0) and well differentiated vs the other groups (F[6,227]=28.4; $P < 0.001$); Cronbach's alpha (internal consistency) was a satisfactory 0.75; items of sleepiness and cataplexy accounted for 45.5% of total variance; sensitivity/ specificity was 94.1%/93.5% (cutoff 13/14), with PPV of 53.3 and NPV of 99.5; the AUC of ROC analysis was 0.97 (95% CI: 0.95-0.99)

[a]Reference numbers relate to full article; [b]All mean/median ages (ranges; SD) and sex percentages given are for participants with narcolepsy; [c]A representative sample/summary of data (all individual studies are too numerous to list); [d]Not a validation study or review of validation studies.

AUC, area under the curve; CETQ, Cataplexy Emotional Trigger Questionnaire; CI, confidence interval; ESS, Epworth Sleepiness Scale; IH, idiopathic hypersomnia; MSLT, Multiple Sleep Latency Test; MWT, Maintenance of Wakefulness Test; NC, narcolepsy with cataplexy; NNH, non-narcoleptic hypersomnia; NpC, narcolepsy with possible cataplexy; NR, not reported; NSAQ, Narcolepsy Status Assessment Questionnaire; NSSQ, Narcolepsy Symptom Status Questionnaire; OSA, obstructive sleep apnea; ROC, receiver operating characteristic; SD, standard deviation; SDB, sleep disordered breathing; SNS, Swiss Narcolepsy Scale; SSI, Stanford Center for Narcolepsy Sleep Inventory; UNS, Ullanlinna Narcolepsy Scale.